Autoimmune neutropenia

Version: 3.0
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Condition for which IVIg use is in exceptional circumstances only

Specific Conditions	• Autoimmune neutropenia
Indication for IVIg Use	• Severe autoimmune neutropenia unresponsive to treatment with G-CSF • Relapse of severe autoimmune neutropenia in a patient demonstrated to have previously responded to Ig therapy
Level of Evidence	Insufficient data (Category 4a)
Description and Diagnostic Criteria	Autoimmune neutropenia is a rare disorder caused by peripheral destruction of autoantibody-sensitised neutrophils by the reticuloendothelial system. While autoantibodies to neutrophil surface antigens may be present they are not specific and do not need to be demonstrated.
Justification for Evidence Category	First line treatment of autoimmune neutropenia is Granulocyte colony-stimulating factor (G-CSF) and antibiotics to treat any infection. In patients with severe infection, there are multiple small case reports and case series supporting the use of Ig therapy (Bux et al, 1991 and Bux et al, 1998 and Getta et al, 2015). Corticosteroids, cytotoxic and immunosuppressant medication and rarely, splenectomy have also been described (Shastri & Logue, 1993 and Capsoni et al, 2005 and Bux et al, 1998). Although some long lasting responses have occasionally been reported following Ig therapy, the overall response is generally short lived and providing an opportunity to treat severe underlying infection.
Diagnosis Requirements	A diagnosis must be made by a Haematologist.
Qualifying Criteria for IVIg Therapy	Severe autoimmune neutropenia unresponsive to treatment with G-CSF This indication should be used for new patients and those that have never received Ig therapy for this condition. For responding patients who have relapsed following Ig therapy please use the indication: **Relapse of severe autoimmune neutropenia in a patient demonstrated to have previously responded to Ig therapy**. • Persistent severe autoimmune neutropenia with a neutrophil count less than 0.5 x10^9/L AND • Recurrent or severe bacterial infection(s) in the last six months AND • Failure to respond to G-CSF treatment AND • Non responsive to other immunosuppressant therapy OR • Other immunosuppressant therapy is contraindicated or has resulted in unacceptable side effects or significant toxicity

Relapse of severe autoimmune neutropenia in a patient demonstrated to have previously responded to Ig therapy

This indication should be used for responding severe autoimmune neutropenia patients who have relapsed following immunoglobulin therapy. For new patients and those that have never received Ig therapy, please use the indication: **Severe autoimmune neutropenia unresponsive to treatment with G-CSF.**

- Persistent severe autoimmune neutropenia

AND

- Recurrent or severe bacterial infection(s) in the last six months

AND

- Previous response following four weeks of Ig therapy demonstrated by improvement in neutrophil count (greater than $0.5x10^9$/L) or a reduction in infections

  AND

- Previous response following four weeks of Ig therapy demonstrated by a reduction in ongoing infections

Review Criteria for Assessing the Effectiveness of IVIg Use

Severe autoimmune neutropenia unresponsive to treatment with G-CSF

Review is not mandated for this indication however the following criteria may be useful in assessing the effectiveness of Ig therapy.

- Improvement in neutrophil count compared to the qualifying assessment

  AND

- Reduction in ongoing infections compared to the qualifying assessment

Relapse of severe autoimmune neutropenia in a patient demonstrated to have previously responded to Ig therapy

Review is not mandated for this indication however the following criteria may be useful in assessing the effectiveness of Ig therapy.

- Improvement in neutrophil count compared to the qualifying assessment

  AND

- Reduction in ongoing infections compared to the qualifying assessment

Dose	Severe autoimmune neutropenia unresponsive to treatment with G-CSF • **Dose** - Up to 2 g/kg in a single or divided dose weekly for 4 weeks The aim should be to use the lowest dose possible that achieves the appropriate clinical outcome for each patient. Refer to the current product information sheet for further information on dose, administration and contraindications. Relapse of severe autoimmune neutropenia in a patient demonstrated to have previously responded to Ig therapy • **Relapse dose** - Up to 2 g/kg in a single or divided dose weekly for four weeks The aim should be to use the lowest dose possible that achieves the appropriate clinical outcome for each patient. Refer to the current product information sheet for further information on dose, administration and contraindications.

Bibliography

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